

# Valvular Heart Disease

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## 50th Anniversary Historical Article

### The Natural History of Aortic and Mitral Valve Disease

by Rapaport (2)

#### ABSTRACT

*An appreciation of the natural history of acquired valvular heart disease is a prerequisite to an understanding of how surgical intervention has altered the natural outlook. The prognosis for a patient with valvular heart disease treated medically is dependent on the stage of the disease at which he is first seen. Therefore, assessment for surgery requires evaluation of the pathophysiologic consequences that have resulted from the hemodynamic alterations. Survival statistics for patients seen at the University of California Medical Center at San Francisco are presented and compared with the data of others. Stenotic lesions appear to have a poorer prognosis than chronic regurgitant lesions and generally warrant surgical intervention at an earlier functional stage of the disease. However, valvular insufficiency produced acutely is poorly tolerated and many constitute a surgical emergency.*

*The selection of patients with mitral or aortic valve disease for surgery obviously requires an appreciation of the natural history of valvular heart disease. One must weigh the expected surgical morbidity and mortality against the anticipated outcome of medical management alone. Therefore, one must know not only the early and late results of surgical procedures for various valvular lesions, but also the pathophysiologic consequences of valvular heart disease at various stages in its natural history. Simple awareness of average survival statistics is inadequate. For example, the prognosis of asymptomatic mitral stenosis differs greatly from that of mitral stenosis that has led to severe pulmonary vascular disease with resultant pulmonary hypertension and cardiac decompensation. The purpose of this review is to examine the major valvular lesions in terms of their pathophysiologic consequences and their natural history in the absence of surgical intervention.*

Originally published in *American Journal of Cardiology*, February 1975.

#### Review

Advances in the evaluation and management of patients with valvular heart disease represent a true twentieth-century

success story. The mortality and morbidity rates in the first half of this century associated with diseases of the aortic and mitral valves were little changed from those of the previous 2,000 years. As recently as 1947 in the third edition of *Heart Disease* (1), Dr. Paul Dudley White indicated that "there is no specific treatment for mitral valve disease" and "there is no treatment for aortic valve disease." The remarkable progress that has been achieved in the last three decades, in terms of pathophysiologic understanding, diagnostic capabilities and corrective surgical and catheter-based techniques, has transformed aortic and

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### INTRODUCTION

In this edition of the *Journal*, we release the ninth in a series of reviews of influential articles that have been previously published in ACC journals, including the *American Journal of Cardiology* (from 1958 to 1982) and *JACC* (from 1983 to the present). The publication of these articles is only one aspect of the ACC's 50th anniversary commemoration, which highlights 50 years of leadership in cardiovascular care and education. The articles are intended to encourage reflection on the remarkable progress made in cardiovascular medicine over time, as well as to acknowledge the amazing prescience of some early investigators in anticipating and, in many cases, later guiding developments in their field.

The working group responsible for selecting these articles and asking reviewers to write editorials solicited suggestions from the ACC's clinical committees and individual members.

The group achieved consensus fairly easily, including whom the group should ask to prepare the accompanying editorials. We initially drew up a list of 14 general areas to cover in this series, but later found that there are several major areas of modern cardiology, prominently molecular cardiology, in which the truly landmark articles have, alas, not yet been published in *JACC*. Therefore, the working group decided not to categorize by subject, but instead, to concentrate on the most important articles.

The working group, a task force of the Subcommittee for the Commemoration of the ACC 50th Anniversary, owes a great deal to Ms. May A. Roustom and the efficient and tireless staff at Heart House for facilitating this project. We also wish to thank all who suggested articles and, most important, the authors who prepared reviews for their willingness to contribute their time and wisdom.

#### *Influential Articles in JACC Working Group*

Sharon A. Hunt, M.D., F.A.C.C.  
Rick A. Nishimura, M.D., F.A.C.C.  
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mitral valve disease from incurable ailments to conditions that are now recognized, characterized and treated routinely by cardiovascular physicians and surgeons, often providing patients with the promise of a normal lifespan and quality of life.

Dr. Elliott Rapaport's report (2) describing the natural history of aortic and mitral valve disease, highlighted in this historical review, was published in 1975, on the threshold of these advances, at a critical transition point in the evolution of treatment of valvular heart disease. In the years preceding Dr. Rapaport's report, the only tools available to evaluate patients and characterize the severity of heart valve disease, and hence the only criteria that could be used to describe their natural history, consisted of symptoms, physical examination, the electrocardiogram, chest X-ray film and, in very selected patients, angiographic and hemodynamic measurements. At that time, surgical treatment was also at a turning point. The earliest model prosthetic ball valves had been implanted for roughly 10 years, and the era of bioprosthetic valves had just begun. The enormous progress in the quarter century since Dr. Rapaport's report includes the appreciation of the importance of ventricular function in determining natural history and outcome; the development of M-mode and two-dimensional echocardiography to assess valve pathology, chamber size and ventricular function; Doppler echocardiography to evaluate the severity of stenotic and regurgitant lesions and pulmonary artery pressures; radionuclide ventriculography to assess ventricular function at rest and with exercise; percutaneous mitral balloon valvotomy as an effective treatment for mitral stenosis; use of blood cardioplegia and retrograde delivery of cardioplegia for intraoperative myocardial protection; bileaflet mechanical valves; stentless bioprosthetic valves, homograft valves and autograft valves for aortic valve replacement; mitral valve repair and chordal sparing mitral valve replacement to maintain integrity of the mitral apparatus in patients with mitral regurgitation; and combined valve replacement/repair and coronary artery bypass graft surgery in patients with concomitant coronary artery and valvular heart disease. In reviewing the previous natural history data leading up to his own natural history study, Dr. Rapaport's paper also represents an important link between the presurgical era of the first half of this century, when there was no effective treatment for aortic or mitral valve disease, and the current era.

A number of Dr. Rapaport's many insights (2) hold true today. These include the long latent period for both stenotic and regurgitant lesions before the onset of symptoms; the inexorable decline in survival once symptoms develop (which is most aggressive in patients with aortic stenosis); the poor outcome of patients with acute aortic or mitral regurgitation treated medically and the need to consider urgent surgery; and the differences in left ventricular wall stress and the resulting hypertrophic response between patients with chronic aortic regurgitation and those with chronic mitral regurgitation. However, several other aspects of Dr. Rapaport's discussion are now less applicable because of the aging of the population, changes in etiology of valve

disease in the developed countries of the world and, importantly, advances in diagnostic and surgical techniques. For example, the predominant cause of aortic stenosis in the U.S. is now degenerative calcific disease in middle-aged and elderly patients rather than congenital bicuspid disease (3), and aortic regurgitation also arises more frequently from a degenerative process than from congenital defects (4). Similarly, the predominant cause of mitral regurgitation is now mitral valve prolapse rather than rheumatic heart disease (4). Although rheumatic heart disease continues to be the cause of virtually all cases of mitral stenosis in adults, its natural course in the U.S. and Canada is now less virulent than it was in the early decades of this century; it is now milder and more delayed (5). It is not uncommon for symptoms of mitral stenosis to first present in middle age, and up to one-third of patients undergoing mitral balloon valvotomy are older than age 65 years (6). In contrast, in the presurgical era, the average age at death was 48 years (7), as noted by Dr. Rapaport.

In addition to the evolution in etiology and natural history, data that are now acquired routinely to characterize virtually every patient with valvular heart disease were not available in 1975. As a result, a different perspective on the natural history of valvular heart disease has emerged over the 25 years since publication of Dr. Rapaport's article.

It is now apparent that for any valve lesion, survival is influenced importantly by age, severity of symptoms, severity of the valvular lesion itself, left or right ventricular systolic function and the presence or absence of concomitant coronary artery disease (4). Additional factors include atrial fibrillation and pulmonary hypertension in mitral valve disease, degree of left ventricular dilation in mitral or aortic regurgitation and severity of left ventricular hypertrophy in aortic stenosis or regurgitation (4). Although there are high-risk subgroups of patients with aortic or mitral valve disease in the current era in whom the natural history without surgical intervention approaches the ominous survival curves reported in Dr. Rapaport's series, this is clearly not the case for asymptomatic patients with normal left and right ventricular size and function.

In addition to the refinement of noninvasive methods for effective risk stratification and identification of patients, symptomatic and asymptomatic, who progress into higher risk subgroups, surgical intervention has also evolved dramatically in the last quarter century. Rather than operating only on the sickest patients with the most advanced forms of valvular heart disease, in whom the high short-term and long-term risks of the operation could be justified, valve replacement or repair is now safely performed at much earlier stages of the natural course of the disease process, often in asymptomatic patients, with excellent long-term results. Early intervention combined with advances in surgery have completely transformed the outlook of patients with aortic or mitral valve disease, compared to that depicted 25 years ago.

Nonetheless, one of the most fundamental aspects of medical decision making in patients with aortic and mitral

valve disease has not changed since 1975. Valve replacement or repair is still not a curative process, and patients who have undergone operation have replaced one set of problematic and serious conditions with another (8). The goal is to operate late enough in the natural course to justify the risks of intervention, but early enough to prevent irreversible ventricular dysfunction, pulmonary hypertension or chronic arrhythmias, or a combination of these complications. Although the balance between the risks of the natural course of the disease and the risks of surgery now often favors early intervention, one must continue to weigh the anticipated early and late outcome of surgical procedures against the expected outcome of medical management alone (2). This decision requires both objective data and sound clinical judgment. This lesson from Dr. Rapaport still holds very true today.

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